CASE REPORT

1:1 atrioventricular conduction in congenital complete heart block

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Abstract

A female neonate with congenital complete heart block developed atrioventricular conduction through an accessory pathway. Despite sinus rhythm and an adequate heart rate she developed severe dilated cardiomyopathy and died at age 14 months. This case illustrates that underlying heart block can be present in individuals with asymptomatic Wolff-Parkinson-White syndrome and that the dilated cardiomyopathy that occasionally accompanies autoimmune congenital heart block is not primarily caused by bradycardia.

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Keywords: congenital heart disease; complete heart block; atrioventricular conduction; Wolff-Parkinson-White syndrome

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Complete recovery of atrioventricular conduction through the atrioventricular node has not been described in a patient with autoimmune congenital complete heart block. We report the case of a female infant with autoimmune congenital heart block who developed 1:1 atrioventricular conduction through an accessory pathway, but subsequently developed severe dilated cardiomyopathy and died at the age of 14 months.

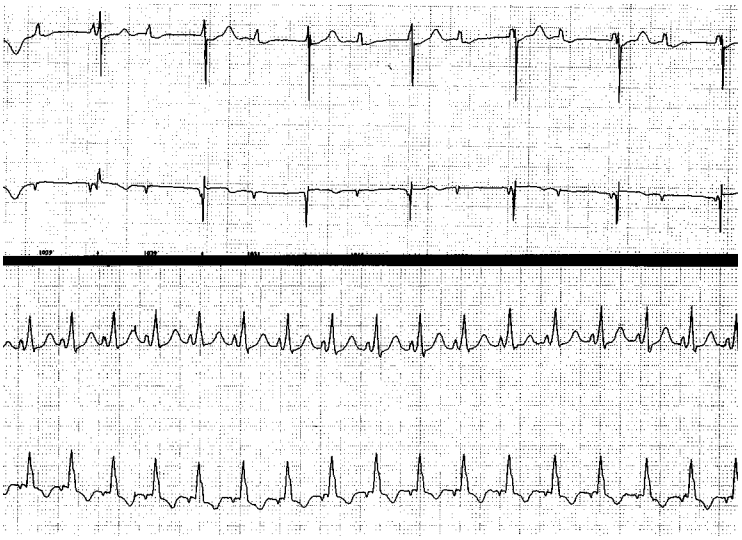

Figure 1 Electrocardiographic tracings from a 24 hour Holter monitor showing variation between complete atrioventricular block and conduction down the accessory pathway.

Case report

Complete heart block was first noted at 24 weeks' gestation. The mother had rheumatoid arthritis and was seropositive for anti-Ro and anti-La antibodies. Oral aspirin was started at 27 weeks' gestation. The baby was delivered at 38 weeks by elective caesarian section. Following delivery, the baby had complete atrioventricular block with a narrow complex ventricular escape rate of 45 beats/min. Left ventricular size and function were within normal limits for age. Implantation of a permanent pacemaker was initially considered, but on the fourth day of life her heart rate suddenly increased to 140 beats/min. Electrocardiography revealed sinus rhythm with a short PR interval and preexcitation, typical of a right sided accessory pathway. A 24 hour Holter recording showed sinus rhythm with conduction down an accessory pathway for approximately 50% of the day with complete heart block for the remainder (fig 1).

The infant initially made good progress, but from the age of 3 months she began to develop signs of heart failure. Echocardiography showed a dilated left ventricle with impaired function. Twelve lead electrocardiography and 24 hour Holter monitoring showed sinus rhythm with conduction down the accessory pathway. Captopril was started with some clinical improvement, but over the following months the left ventricle continued to dilate with worsening function.

At age 9 months, conduction in the accessory pathway failed and she became bradycardic and lethargic. Electrocardiography showed complete atrioventricular block with a ventricular rate of 40 beats/min. A permanent single chamber endocardial pacemaker was implanted. Aortic root angiography confirmed normal coronary arteries. Myocardial biopsy specimens showed mild cellular hypertrophy with no inflammatory cells and no fibrosis.

Two months following pacemaker implantation, she was readmitted with worsening heart failure. The left ventricle was severely dilated with poor function. Five days following admission, she suddenly deteriorated with low cardiac output and died.

Discussion

There has been much debate recently about whether to ablate individuals with asympto-

matic Wolff-Parkinson-White syndrome (WPW).[1] In most infants with isolated congenital complete heart block it is believed that antibodies to soluble tissue RNA cross the placenta and damage conducting tissue.[2 3] Our patient demonstrates that the atrioventricular node may be damaged while accessory conducting tissue is unaffected. It is possible, therefore, that some individuals with asymptomatic WPW will have underlying complete heart block, which will only become apparent following ablation. Clearly in any patient with asymptomatic WPW, function of the atrioventricular node should be confirmed before ablation, and if this is not achieved perhaps the procedure should be reconsidered. It could be argued that if there is underlying heart block an accessory pathway is an unreliable form of conduction, and that such patients should be identified. Atrial fibrillation degenerating to ventricular fibrillation is thought to be the main reason for sudden death in WPW,[4 5] however, it is possible that sudden loss of accessory pathway conduction where there is underlying complete heart block could also be a cause.

It is becoming increasingly recognised that dilated cardiomyopathy is a rare but serious outcome of autoimmune congenital complete heart block.[6] The cause of the cardiomyopathy is unclear but may be related to an autoimmune myocarditis occurring in utero rather than primarily caused by bradycardia. Our patient supports this theory as she developed a severe dilated cardiomyopathy despite a normal sinus rate with atrioventricular synchrony. Fetal hydrops develops in a small percentage of cases with congenital heart block and is believed to be secondary to autoimmune myocarditis rather than primarily caused by bradycardia.[7] There has been some success in reducing hydrops by treating the mother with steroids.[7] Our patient's mother did receive aspirin from 27 weeks' gestation, but neither heart block nor cardiomyopathy was prevented. Perhaps there is an argument for giving maternal steroids in all cases of fetal heart block in the hope that the serious, albeit rare, complication of cardiomyopathy might be prevented.

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